

Complete Listing of Claims Pursuant to 37 C.F.R. §1.121

Pursuant to 37 C.F.R. §1.121 the following is a complete listing of the claims of the present application. In this set of claims, please amend the claims by canceling claims 1-9 and adding the following new claims: With the amendments to the aforementioned claims, the following listing of claims will replace all prior versions, and listings, of claims in the application:

1-9 [Cancelled]

10. [Currently Amended] A method of treating lysosomal storage diseases ~~in a subject in need thereof, said method in an animal~~ comprising:

administering to said ~~subject animal~~ a compound of claim 14 comprising RAP or RAP polypeptide conjugated to a therapeutic agent, wherein said compound crosses the cell membrane, enters cells and is delivered to lysosomes in an amount effective to treat alleviate one or more symptoms of said lysosomal storage disease.

11. [Withdrawn] The method of claim 10, wherein the agent is an enzyme deficient in the lysosomal storage disease.

12. [Withdrawn] The method of claim 11, wherein the lysosomal storage disease is selected from the group consisting of aspartylglucosaminuria, cholesterol ester storage disease, Wolman disease, cystinosis, Danon disease, Fabry disease, Farber lipogranulomatosis, Farber disease, fucosidosis, galactosialidosis types I/II, Gaucher disease types I/II/III, Gaucher disease, globoid cell leukodystrophy, Krabbe disease, glycogen storage disease II, Pompe disease, GM1-gangliosidosis types I/II/III, GM2-gangliosidosis type I, Tay Sachs disease, GM2-gangliosidosis type II, Sandhoff disease, GM2-gangliosidosis, α -mannosidosis types I/II, β -mannosidosis, metachromatic leukodystrophy, mucolipidosis type

I, sialidosis types I/II mucolipidosis types II /III I-cell disease, mucolipidosis type IIIC
pseudo-Hurler polydystrophy, mucopolysaccharidosis type I, mucopolysaccharidosis type II,
Hunter syndrome, mucopolysaccharidosis type IIIA, Sanfilippo syndrome,
mucopolysaccharidosis type IIIB, mucopolysaccharidosis type IIIC, mucopolysaccharidosis
type IIID, mucopolysaccharidosis type IVA, Morquio syndrome, mucopolysaccharidosis type
IVB Morquio syndrome, mucopolysaccharidosis type VI, mucopolysaccharidosis type VII,
Sly syndrome, mucopolysaccharidosis type IX, multiple sulfatase deficiency, neuronal
ceroid lipofuscinosis, CLN1 Batten disease, Niemann-Pick disease types A/B, Niemann-Pick
disease, Niemann-Pick disease type C1, Niemann-Pick disease type C2, pycnodynatosostosis,
Schindler disease types I/II, Schindler disease, and sialic acid storage disease.

13. [Withdrawn] The method of claim 10, wherein the agent is selected
from the group consisting of aspartylglucosaminidase, acid lipase, cysteine transporter,
Lamp-2, α -galactosidase A, acid ceramidase, α -L-fucosidase, β -hexosaminidase A, GM2-
activator deficiency, α -D-mannosidase, β -D-mannosidase, arylsulfatase A, saposin B,
neuraminidase, α -N-acetylglucosaminidase phosphotransferase, phosphotransferase γ -
subunit, L-iduronidase, iduronate-2-sulfatase, heparan-N-sulfatase, α -N-
acetylglucosaminidase, acetylCoA:N-acetyltransferase, N-acetylglucosamine 6-sulfatase,
galactose 6-sulfatase, β -galactosidase, N-acetylgalactosamine 4-sulfatase,
hyaluronoglucosaminidase, multiple sulfatases, palmitoyl protein thioesterase, tripeptidyl
peptidase I, acid sphingomyelinase, cholesterol trafficking, cathepsin K, α -galactosidase B,
and sialic acid transporter.

14. [Currently amended] A compound comprising Receptor-Associated
Protein (RAP) conjugated to an enzyme that is deficient in [[the]] a lysosomal storage
disease.

15. [Previously presented] The compound of claim 14, wherein said
enzyme is selected from the group consisting of aspartylglucosaminidase, acid lipase,

cysteine transporter, Lamp-2, α -galactosidase A, acid ceramidase, α -L-fucosidase, β -hexosaminidase A, GM2-activator deficiency, α -D-mannosidase, β -D-mannosidase, arylsulfatase A, saposin B, neuraminidase, α -N-acetylglucosaminidase phosphotransferase, phosphotransferase γ -subunit, L-iduronidase, iduronate-2-sulfatase, heparan-N-sulfatase, α -N-acetylglucosaminidase, acetylCoA:N-acetyltransferase, N-acetylglucosamine 6-sulfatase, galactose 6-sulfatase, β -galactosidase, N-acetylgalactosamine 4-sulfatase, hyaluronoglucosaminidase, multiple sulfatases, palmitoyl protein thioesterase, tripeptidyl peptidase I, acid sphingomyelinase, cholesterol trafficking, cathepsin K, α -galactosidase B, and sialic acid transporter.

16. [Withdrawn] The compound of claim 14, wherein said enzyme deficient in the lysosomal storage disease is human α -L-iduronidase.

17. [Previously presented] The compound of claim 14, wherein said enzyme deficient in the lysosomal storage disease is human α -glucosidase.

18. [Previously presented] The compound of claim 14, wherein said RAP is directly covalently linked to said enzyme.

19. [Previously presented] The compound of claim 14, wherein said RAP is linked to said enzyme through a linker.

20. [Previously presented] The compound of claim 19, wherein said linker is an amino acid linker of between about 5 to about 50 amino acids.

21. [Previously presented] The compound of claim 14, wherein either said RAP or said enzyme is derivatized with a polyethylene glycol moiety.

22. [Previously presented] A composition comprising the compound of any of claims 14 to 21 and a pharmaceutically acceptable carrier.

23. [Previously presented] The composition of claim 22, wherein said composition is prepared as an aerosol formulation to be administered via inhalation.

24. [Previously presented] The composition of claim 22, wherein said composition is prepared for administration intravenously.